

# **Palliative Care for an Infant with Short Bowel Syndrome and Advanced Liver Disease**

## **Several physicians offer commentary on when it may be ethical to choose to not pursue life-prolonging treatments for terminally ill infants.**

Commentary by Mark Sheldon, PhD, and by Roytesa Savage, MD, Ronald M. Perkin, MD, MA, and Joseph R. Zanga, MD

After 5 months of routine NICU Care, and treatment for malabsorption and malnutrition, Mary was discharged from the NICU and allowed to go home with her parents. Eight hours after discharge, her parents brought her back to the hospital with fever and vomiting. During this second hospitalization, Mary had bacterial and fungal infections, multiple changes in her vascular access sites, and complications, including advanced liver disease, from the total parenteral nutrition (TPN). Her liver dysfunction was characterized by abnormal coagulation, only partially corrected by blood products and vitamin K, hypoalbuminemia, and hypoproteinemia. She bled from her nose and mouth after crying or sneezing, and extensively from her ostomy site. Mary had multiple episodes of hypovolemic shock that required blood transfusions. Her massive hepatosplenomegaly interfered with respiration.

Early in her NICU stay Mary's physicians discussed with the Janes the possibility of transporting Mary to another medical center for an intestinal transplant. Mary's parents appeared to understand the seriousness of their daughter's condition and wanted the doctors to do "everything possible" for her. Mary's liver dysfunction progressed and she became more edematous, had skin breakdown, and had to be more frequently volume resuscitated and transfused. Her tenuous condition now made it impossible to consider moving her to another location for a transplant. Mary's physicians considered her condition terminal and could see that she was suffering.

Dr. Andrews and her colleagues tried to talk to the Janes about palliative care and the imminent death of their daughter. In one instance Dr. Andrews approached Mrs. Jane, who never left the hospital unless her husband or mother came to relieve her, but Mrs. Jane stopped Dr. Andrews in mid-sentence.

"I see where you're going with this, Dr. Andrews, but my baby is strong. Children are resilient. Mary got well enough to go home once and she'll do it again, we've just got to give her a little time."

\*The patient's name has been changed to protect her privacy and that of her parents.

### **Commentary 1**

by Mark Sheldon, PhD

This is a case where it is important for Dr. Andrews to remember 2 things: who the patient is and the Hippocratic Oath. The patient is Mary Jane and the Hippocratic Oath states, "...I will keep them [patients] from harm."

It is particularly difficult to remember who the patient is when the patient is a child and there are loving parents involved. Psychologists indicate that there is hardly an experience more painful and traumatic than the loss of a child. Parents clearly and reasonably expect to die before their children; a child's death goes against the natural order. The

essential role of parent is to protect the child. Furthermore, if the physician is a parent, he or she may identify too strongly with the parents, or may feel guilt in connection with the fact that his or her own child is healthy while the patient is dying. All of this means that the tendency of the physician will be to continue to treat as long as there are treatment alternatives to pursue.

Much has been written about the importance of recognizing that the family is often centrally involved in the treatment of any patient and may be even more involved in the cases of pediatric patients. No one would take issue with a concern to accommodate the wishes of the family to the extent possible. For physicians and staff the hospital world is routine, and they may not always appreciate the challenges families confront when a loved one is hospitalized. Given the extent to which the treatment of illness and disease is so completely institutionalized in our society, however, it is not unusual for families to face, for the first time and all at once, 3 very significant challenges: an exceedingly technologically complex and unfamiliar environment, an experience of great emotional and psychological complexity, and enormous personal responsibility. And it is the task of the physician and staff to educate the family in regard to all 3 and to help the family cope.

The neonatal unit, particularly, is a place of great technological complexity. The case description makes clear that everything has been done for this baby. One imagines that the parents have been engaged in a huge effort to understand the options that the technology has made available for the treatment of their baby, although it is not clear that they have fully appreciated the cost to their baby of such efforts. Considering their baby's pain and suffering, one is not able to imagine the emotional and psychological anguish that the Janes have experienced. It is likely that nothing in their previous lives prepared them for the profound responsibility that they are being asked to shoulder.

Another feature of this case, which Dr. Andrews should take into account, is that the neonatal unit is a place that requires a team effort. Nurses have been involved fundamentally in the care of this infant, probably at great emotional cost to them. As the baby's skin breaks down, they, more than anyone, are aware of what the baby must endure if additional life-saving interventions are undertaken. The physician is accountable to her staff, as well as the parents, and the conscientious staff takes the imperative to do no harm just as seriously as do the physicians.

True, much is remarked about the resilience of children. Pediatricians are often surprised when a child pulls through. But it is clear that there are cases where such an outcome will not be possible, and this is one of those cases. This case has involved the pursuit, by clinicians, of every available heroic intervention that might provide benefit. However, the issue, at every step of the way, has been whether any benefit could be achieved that would outweigh the burden, the discomfort, and the suffering to which the baby was subjected as these interventions were undertaken.

At this point, it is clear that no further benefit can be provided, all options have been exhausted, and the baby stands only to experience more suffering and pain. Furthermore, she will die regardless of what is done. This is the time for Dr. Andrews to bring her professional judgment to bear. This is what being trained to exercise professional judgment is all about. Gently and firmly, she must refuse any interventions that aim at prolonging the baby's life. She must say "no" to the parents and explain that, ultimately, the baby is her patient, her responsibility, and that her professional ethic requires her to refuse to intervene aggressively to prolong this baby's suffering. It is time, she must explain, to shift entirely to palliative care in an effort to keep the baby comfortable and allow her to die. If the parents refuse, she must tell the parents that she can no longer care for this baby, that what they are asking her and her staff to do constitutes a harm that is not acceptable.

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## **Commentary 2**

by Roytesa Savage, MD, Ronald M. Perkin, MD, MA, Joseph Zanga, MD

Mary Jane was indeed a resilient infant, and, over the course of her short life, her parents, particularly her mother, not unexpectedly bonded with her and likely contributed to that resilience. Unfortunately her short bowel syndrome and

advanced liver disease made it impossible to nourish her adequately by either parenteral or enteral means. Intestinal transplant was considered but quickly became an unreasonable option in light of the severity of Mary's liver dysfunction. Even in the best of circumstances, however, likelihood of 1-year survival after intestinal transplant is only approximately 50 percent [1], making the decision to operate a difficult choice for parents and a difficult recommendation for physicians to make.

In this infant's case other choices were equally difficult, and ethical discussions began to assume as much of a role in her care as the medical discussions. Nonmaleficence was the first consideration, given that the total parenteral nutrition, initiated to sustain the child's life, was ultimately hastening her death by destroying her liver. Withholding this therapy was therefore ethically permitted if not obligated [2-3].

The physicians, not the parents, ultimately concluded that the greatest benefit to Mary, and the most appropriate therapy, was to offer comfort care. In this they were supported by the American Academy of Pediatrics' Committee of Bioethics which in 1994 concluded that "continuing non-beneficial treatment harms many patients and may constitute a legal, as well as moral, wrong" [4].

As consideration of the appropriateness of palliative care continued, some members of Mary's health care team wondered whether it might be illegal to withdraw nutrition and hydration, citing the 1984 Department of Health and Human Services amendments to the Child Abuse Prevention and Treatment Act. These regulations, commonly known as the "Baby Doe Regulations," require that, except under certain specified conditions, all newborns receive maximal life-prolonging treatment. While still criticized by some, the Baby Doe Regulations continue to influence decision making for terminally ill newborns [5-6]. There is clear consensus, however, that withdrawal of artificial nutrition and hydration when they are more harmful than helpful and when the child's parents or legal guardians are in agreement, is fully acceptable and justifiable both legally and morally.

For a variety of reasons Mary's parents (and others in similar circumstances) were unwilling to consider palliative care. Health professionals are often unwilling to embark on this course due to concern about hastening death. Ethical consideration of aggressive palliation often includes a discussion of the principle of double effect [7]. This principle, which stems from the moral theology of Thomas Aquinas, states that an action with both a good and bad effect is ethically permissible if the following conditions are met:

1. The action itself is morally good or at least indifferent.
2. Only the good effect is intended (even though the bad or secondary effect may be foreseen).
3. The good effect must not be achieved by way of the bad.
4. The good effect must outweigh the bad.

Often the most difficult aspects of initiating such an approach to care is reluctance on the part of health professionals [8], and the lack of understanding by patients, parents, or relatives. It is of course not a decision to be made lightly, and in the case of Mary Jane it is clear that additional discussion was required between Dr. Andrews and the parents before such an approach could be accepted.

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